

# Mediastinal thymoma in children: A cytological diagnosis

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## ABSTRACT

Mediastinal masses in children comprise a heterogeneous group of malignant and benign neoplasms. The majority of malignant tumors of mediastinum are Hodgkins and non-Hodgkins lymphomas, germ cell tumors, and neuroblastomas. Thymomas, thymic carcinoid, and thymic carcinomas constitute thymic epithelial neoplasms (TEN). Thymoma is a primary tumor of the thymus epithelial cells and one of the most common neoplasms in the anterosuperior mediastinum, especially in middle-aged or older adults. Thymic epithelial neoplasms are usually seen in the latter decades of life and rarely occur in children. We hereby report a case of 13-year-old girl presenting with thymoma.

**Key words:** Childhood thymoma, mediastinum, thymoma

## INTRODUCTION

Thymic epithelial neoplasms are usually seen in the fourth and fifth decades of life and rarely occur in children.<sup>[1]</sup> Thymic tumors account for 1.5% of mediastinal masses in children.<sup>[2]</sup> Thymoma is a primary mediastinal neoplasm arising from or exhibiting differentiation toward thymic epithelial cells, typically with the presence of non-neoplastic lymphocytes.<sup>[3]</sup> The overall incidence of thymoma in the United States is 0.15 per 100,000 person-years.<sup>[4]</sup> Thymomas can be classified as benign or malignant. Cytologically benign thymomas have been classified as being lymphocyte rich, epithelial cell rich, or spindle cell type.<sup>[5]</sup> Fine-needle aspiration cytology plays a significant role in the diagnosis of anterior mediastinum masses, contributing to the early management and choice of the optimal therapeutic manipulation.<sup>[6]</sup> Thymomas are classified as cortical, medullary, or mixed. Thymomas that are cytologically malignant are uncommon. Such tumors usually are squamous cell carcinomas. Other types include

sarcomatoid carcinoma, clear cell carcinoma, basaloid carcinoma, and mucoepidermoid carcinoma.<sup>[5]</sup> Aspirates of thymomas are distinguishable from other lesions, and FNA and CT-guided core needle biopsy are proven methods for investigating mediastinal masses. The cytologic diagnosis of thymoma can be extremely challenging. Here we report a case of thymoma diagnosed on FNA in a 13-year-old female who presented with a mediastinal mass.

## CASE REPORT

A 13-year-old girl presented with complaints of weakness, anorexia, weight loss, intermittent fever, sweating, chest pain, cough, and progressive respiratory distress for the previous 2 months. Physical examination revealed paleness, tachypnea (28/min), suprasternal, and intercostal retractions, and decreased left chest breath sounds. She was offered antituberculous treatment for 6 months.

Laboratory findings revealed anemia (hemoglobin: 6.5 g/dL), high sedimentation rate (94 mm/h), and elevated Lactic acid dehydrogenase (547 U/L). All of her urine analysis, serum biochemical tests, blood gases, Alpha fetoprotein, beta-Human chorionic gonadotropin, and urine vanillylmandelic acid levels were normal. Bone marrow aspiration and peripheral blood smear were normal. Chest X-ray demonstrated a bulky mass in the anterior mediastinum.

Two dimensional, three dimensional, and four dimensional

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ultrasound scanning of the chest revealed a well-outlined soft tissue mass measuring 126 × 104 × 99 mm in left mid and lower thoracic cavity pushing the heart on the right side. The mass was also extending upto the superior mediastinum. The mass consisted of two components, the peripheral soft tissue component showed internal vascularity, the central component showed no internal vascularity and showed hypoechoic fluid contents. Computerized tomography of thorax showed an anterior mediastinal mass of size 12 × 12 × 10 cm. The ultrasonography and computerized tomography of the abdomen and pelvis were normal. Whole body bone scan showed increased uptake in the sternum.

## CYTOLOGICAL FINDINGS

Ultrasound-guided fine-needle aspiration from the mass yielded blood-tinged material. Smears were air dried and stained with giemsa and also fixed in 95% alcohol and stained with papanicolaou stain. Smears showed a mixed population composed of epithelial cells and lymphocytes. The epithelial cells are present in clusters with a little variation in size and shape of the component cells, while the lymphocytes were dispersed among the epithelial cell clusters in a necrotic background. A cytological diagnosis of a lymphoepithelial lesion [Figures 1 and 2] possibly thymoma was offered. A biopsy from mediastinal mass was performed. Histopathological examination revealed features of thymoma (cortical type).

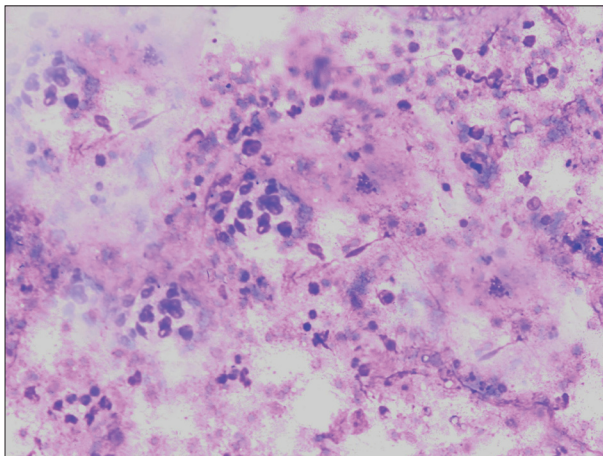
## DISCUSSION

Thymoma is a rare primary tumor of the epithelial cells of thymus and also the most frequent tumor of the anterosuperior mediastinum. Its incidence increases with age (70% of cases are observed in patients past the age of 40). Thymomas are usually asymptomatic or accompanied

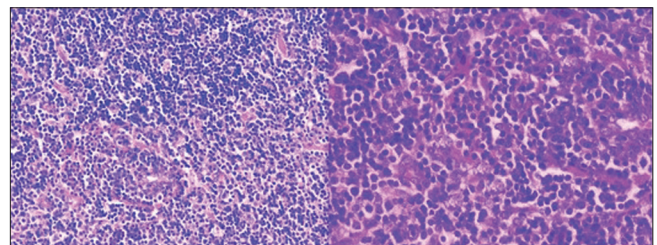
with atypical clinical symptoms such as cough, chest pain, dysphagia, and dyspnoea or may be associated with a wide variation of paraneoplastic syndromes, such as myasthenia gravis, red cell hypoplasia, or pancytopenia. These syndromes may be the first clinical manifestation in an otherwise asymptomatic patient.<sup>[6]</sup> Thymomas can exhibit a spectrum of autoimmune phenomena, comprising neuromuscular, hematopoietic, dermatologic, rheumatic/vasculitic, hepatic, and renal diseases.<sup>[3]</sup> These neoplasms are composed of a mixed population of cells, such as epithelial cells and lymphocytes. Only the epithelial cells show neoplastic features. They may be round, oval, or spindle-shaped and tend to occur in tightly knit clusters. The lymphocytes are predominantly of the T-type, originate in bone marrow and are diffusely distributed. In 40% of cases the tumors are predominantly lymphocytic, in 20% they are mixed and the rest are predominantly of the epithelial type.<sup>[6]</sup> The vast majority of thymomas do not present considerable atypia or anaplasia. In 90% of cases, they are well encapsulated and multilobulated, while in 10% a rupture of the capsule and invasion to the adjacent tissues are observed.

Thymoma should be differentiated from other anterior mediastinal neoplasms with epithelial and/or lymphoid differentiation, including non-Hodgkin (NHL) and Hodgkin lymphomas, thymic carcinomas, and germ-cell malignancies. Limitations of the cytological method include an unproven ability to definitively separate thymoma into specific WHO subtypes using cytology alone, and to determine capsular invasion.<sup>[3]</sup>

The biological behavior of thymomas is unpredictable. Extrathoracic metastases are rare and comprise the cervical lymph nodes and the liver. The stage of the tumor at the time of diagnosis and the adequacy of the surgical excision are among the factors that influence the outcome of these tumors. The presence of clinical symptoms, large tumor size, local invasion or metastases at the time of the surgery, and predominant epithelial features are poor prognostic factors.<sup>[7]</sup> Thymic epithelial carcinomas are rarely seen in childhood. Only two pediatric cases were described in 19 clinopathological studies including 305 cases analyzed by Chung.<sup>[8]</sup> Only 14 cases younger



**Figure 1:** Biphasic pattern of cohesive epithelial cells in a background of lymphoid cells; MGG:X400



**Figure 2:** Scattered plump epithelial cells with vesicular nuclei, distinct nucleoli in a background of lymphocytes-thymoma; (H and E, X200, X400)

than 18 years of age were found in a study conducted by Yaris *et al.* that were either single case reports or existed in case series. Histopathologic definition was available in 12 of 14 cases. The median age of cases was 13 years and there was a male predominance (M/F:9/5). Radiologically, masses may be associated with pleural effusions and involvement of neighboring structures such as pleura and pericardium are frequently observed. A multimodality approach that includes surgery, chemotherapy, and radiation therapy is suggested for treatment. Surgery is the mainstay of the treatment. However, complete resection is generally impossible or not feasible at diagnosis because of local invasion of important structures and metastasis. Radiotherapy improves the local control of the disease. Cisplatin-based regimens have also proved to be effective in the treatment. In cases similar to ours, neoadjuvant chemotherapy was proposed to improve the resectability of the mass.<sup>[2]</sup>

## CONCLUSION

Thymoma is a primary tumor of the thymus epithelial cells and one of the most common neoplasms in the anterosuperior mediastinum. Thymic epithelial neoplasms are usually seen in the fourth and fifth decades of life. Thymomas rarely occur in children. FNA of anterior mediastinal thymic lesions generally yields adequate cellular tissue with distinct cytologic and immunophenotypic features that enables thymoma diagnosis. FNA plays a significant role in

clarifying the nature of these lesions and may contribute to the early management and choice of the optimal therapeutic manipulation. FNAC along with clinical and radiological findings renders a definitive diagnosis of thymoma.

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